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Sarcomatoid carcinoma of the urinary bladder: A case report and short review of the literature

Mesaneğin sarkomatoid karsinomu: Olgu sunumu ve literatürün kısa gözden geçirilmesi

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Abstract

Although the most common malignant formations in the bladder are urothelial carcinomas, there are also tumors with different differentiation degrees like sarcomatoid carcinoma of the urinary bladder. It is a rare and highly aggressive type of tumor and consists of malignant epithelial and sarcomatous components. The literature about the subject is extremely limited, and there is no consensus on treatment protocols. This case report aims to present a case of 71-year-old man who presented with macroscopic hematuria and diagnosed with sarcomatoid carcinoma of the urinary bladder.

Keywords: Sarcomatoid, Carcinoma, Bladder

Öz

Mesaneğin en sık izlenen malign oluşumlar ürotelyal karsinomlar olmakla birlikte sarkomatoid karsinom gibi farklı diferansiyasyon gösteren tümörler de bulunmaktadır. Bu nadir izlenen tümöral yapılar malign epitelyal ve sarkomatöz komponentler içeren oldukça agresif davranışlı oluşumlardır. Bu konu ile ilgili literatür bilgileri son derece sınırlı olup tedavi protokolleri hakkında fikir birliğine varılan bir rejim mevcut değildir. Bu olgu sunumunda makroskopik hematüri ile başvuran ve mesaneğin sarkomatoid karsinomu tanısı alan 71 yaşındaki erkek hastanın literatür bilgileri eşliğinde tartışılması amaçlanmıştır.

Anahtar kelimeler: Sarkomatoid, Karsinom, Mesane

Introduction

Bladder cancer is a significant health problem with increasing incidence in parallel with the growing prevalence of tobacco smoking worldwide. It is the ninth most common cancer in the world and ranks second among all types of cancer in the urinary tract. Besides, clinical studies showed that more than 100,000 patients were diagnosed with a muscle-invasive or advanced disease each year around the globe [1,2]. The majority of bladder tumors (95-98%) consist of transitional cell (urothelial) cancers [2]. There are also some rare malignant tumors such as hemangiopericytoma, leiomyosarcoma, liposarcoma, rhabdomyosarcoma, small cell bladder cancer, angiosarcoma, chondrosarcoma, malignant fibrosis histiocytoma, malignant peripheral nerve sheath tumor, sarcoma, and primitive neuroectodermal tumor in the bladder [3].

Sarcomatoid carcinoma of the urinary bladder accounts for approximately 0.3% of all bladder tumors [4]. It is defined by the World Health Organization as a biphasic tumor consisting of malignant epithelial and mesenchymal components [5]. More than 70% of the diagnosed patients are advanced-stage, and the most common presenting complaint is hematuria. Compared with conventional urothelial carcinomas in the bladder, it appears as a cancer with rather a poor prognosis showing more invasive and aggressive behavior [6].

A literature search showed that there are a very limited number of publications on sarcomatoid cancer of the bladder, as opposed to the common malignant formations of the bladder. Herein, we aimed to present a case with bladder cancer whose pathological results indicated sarcomatoid carcinoma.

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Case presentation

A 71-year-old male patient presented with a 1-year history of occasional macroscopic hematuria. A detailed history revealed that he had been smoking three packs a day for 40 years. Besides, he had undergone transurethral resection due to a bladder tumor at another hospital about ten years ago and yet he did not attend follow-ups. The operative and pathological details were not available. He had no other history of surgery or a systemic disease. On physical examination, the only pathological finding was suprapubic sensitivity. His laboratory test results were as follows: creatinine 1.31 mg/dl; urea 47.45 mg /dl; hemoglobin 7.11 g/dl.

The patient was hospitalized and three units of blood transfused. Computed tomography revealed a heterogeneously enhancing mass which boundaries cannot be selected clearly and reaching the widest 160mm x102 mm dimensions and covering the bladder lumen almost completely. It was considered as bladder tumor and tumor-associated hematoma. The perivesical fat tissue was normal. Imaging procedure showed no additional major pathological finding (Figure 1).



Figure 1: Computed tomography scan showing heterogeneously enhancing bladder mass with hematoma (white arrows)

Bladder irrigation was performed through catheterization and endourological examination was carried out under general anesthesia. Pre-cystoscopy bimanual palpation was unremarkable. Cystoscopy revealed hematoma filling the bladder lumen almost completely, and a broad-based solid tumor with necrotic material on the surface occupying the trigon and right wall of the bladder. Transurethral resection was performed. Surgical pathology report revealed that sarcomatoid carcinoma was present, and the muscularis propria was positive for tumor. Then the patient underwent radical cystectomy and ileal loop diversion. The microscopic evaluation of tissue samples demonstrated that the tumor was composed of epithelioid cells with irregular vesicular nuclei and pale cytoplasm with indistinct borders. Some cells showed spindle morphology with bipolar nuclei and formed bundles with storiform-like growth pattern. The tumor infiltrated both the detrusor muscle and perivesical fat tissue (Figure 2). Immunohistochemical analysis revealed bladder mucosa to be the primary site of origin (Figure 3).

The patient was followed up in the intensive care unit due to respiratory disorder postoperatively and died of respiratory failure on postoperative day three.

Our study was conducted in accordance with the Helsinki Declaration Principles, and was performed after patient's approval with informed consent.

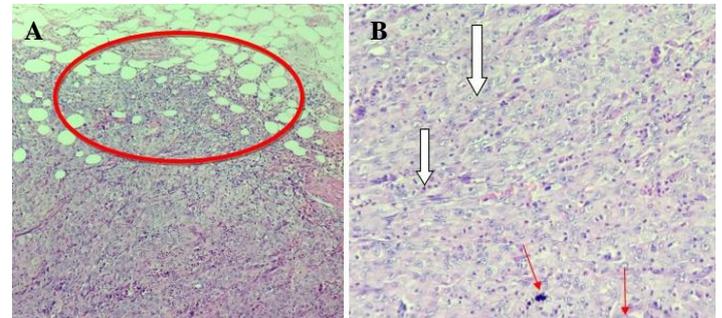


Figure 2: Histopathological examination of the bladder mass, A: Tumoral invasion through muscularis propria to the perivesical fat tissue (red ellipse) (x10), B: Semi epithelioid-spindle cells arranged in trabeculae and bundles (white arrows), frequent mitoses and highly variable nuclear atypia (red arrows) (x20)

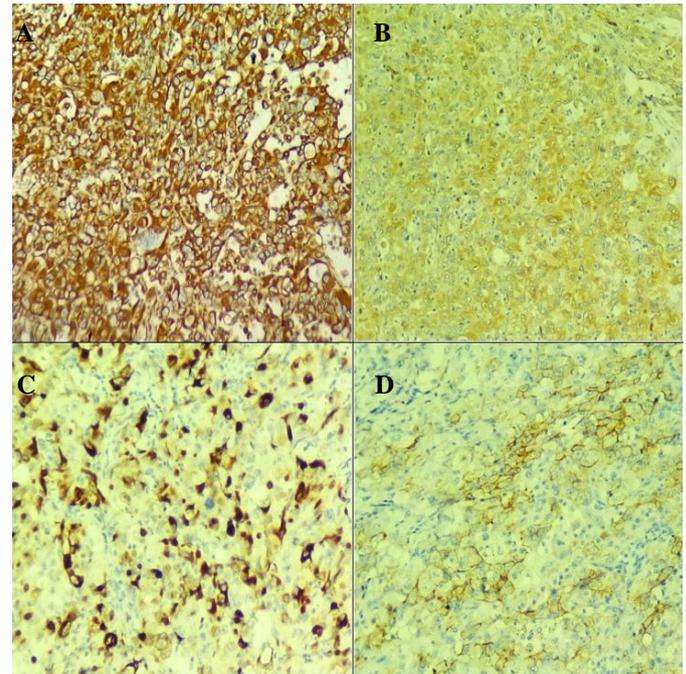


Figure 3: Immunohistochemical analyses of the bladder mass: Vimentin (A), Fascin (B), CK7 (C) and EMA (D) expressions in tumor cells

Discussion

Sarcomatoid carcinoma can be seen in many different parts of the body such as the genital system, thymus, skin, breast, spleen, pancreas, peritoneum and upper and lower respiratory system. However, the most common organ involved is the uterus in postmenopausal women, and the bladder in men [4,7]. The location of sarcomatoid cancers is also important in the clinical course of the tumor. Those seen in the upper respiratory tract or gastrointestinal system have a rather better prognosis. However, the tumors diagnosed in the urogenital system, such as the bladder, show a highly aggressive course [4]. Sarcomatoid cancers are seen most commonly in individuals over 60 years of age and less frequently in women than men. In previous publications, sarcomatoid cancers of the bladder have been described using a variety of terminologies such as carcinosarcoma, metaplastic cancers, Müllerian tumors, spindle cell cancer, heterologous differentiated tumors or malignant mixed mesodermal tumors [4,8].

Symptomatology in sarcomatoid cancers of the bladder is very similar to the transitional cell tumors frequently observed in the bladder. As the macroscopic hematuria is the main complaint, the recurrent urinary tract infections, suprapubic pain, and dysuria are the other signs and symptoms [3,9]. Similarly, the main symptom was macroscopic hematuria in our case. There

are many factors strongly associated with sarcomatoid cancer of the bladder such as smoking, radiation exposure, cyclophosphamide use, and recurrent cystitis [6,10]. The patient discussed in this report had a history of heavy smoking. Previous studies showed that tumor diameters were highly variable. The most common site of tumor cells is in the bladder base, and most of them are exophytic, ulcerated, invasive in-nature with bleeding and necrotic areas [4,11]. Pathological evaluation revealed that the tumor consists of transitional epithelial cells as well as sarcomatoid components. The sarcomatoid component comprises a mixture of a spindle or round pleomorphic cells resembling those seen in leiomyosarcoma, chondrosarcoma or other sarcomatoid neoplasms. The carcinomatous component may show a papillary or non-papillary configuration with features of adenocarcinoma, squamous cell carcinoma or small cell carcinoma; or may be present only as carcinoma in situ. The tumor may contain myxoid or sclerosing areas, as well as necrosis [12].

Perret et al. [13] analyzed 47 patients with sarcomatoid cancers of the bladder and concluded that 83% of them had muscularis propria involvement. Erdemir et al. [4] evaluated 159 patients who underwent radical cystectomy in their department and found that only 0.031% of them had sarcomatoid cancer. They also reported that all patients diagnosed with sarcomatoid cancer were in stage 2 and above. In a similar study, Beltran et al. [10] found that all patients evaluated in their clinical analysis were in advanced stages. Similarly, the case in our study was in the advanced stage.

There are several treatments for sarcomatoid cancers of the bladder. These are transurethral tumor resection, partial cystectomy, radical cystectomy, and radiotherapy and/or chemotherapy protocols applied with these treatment modalities. However, all these treatment options have highly limited efficacy [4,14]. The survival time is quite short, below 24 months after surgery [4]. A retrospective study involving 221 cases with sarcomatoid cancers of the bladder reported that the 5-year cancer-specific survival rate after radical cystectomy was 20.3%. In the same study 1, 5 and 10 year survival rates were reported to be 53.9%, 28.4% and 25.8%, respectively [15]. In another study evaluating patients with the diagnosis of sarcomatoid cancers of the bladder, Wright et al. [6] performed transurethral resection in 119 patients and cystectomy in 79 patients, and a total of 34 patients received postoperative radiotherapy. The researchers reported the mean survival time was 14 months. In our study, the patient died of postoperative respiratory failure. Therefore, we do not have disease-related survival data.

Transurethral resection is a very critical procedure for diagnostic and therapeutic purposes in patients with an image suggesting a mass within the bladder lumen. With this endourological intervention applied in almost all urology clinics, the type and extent of the cancer cells and the prognosis are determined, and treatment of the disease is planned. The patient in our report had presented with macroscopic hematuria. The first invasive procedure he underwent was cystourethroscopy followed by transurethral resection. There is no standardized approach to the treatment of sarcomatoid cancers of the bladder. However, many urology units carry out radical cystectomy and then chemotherapy or radiotherapy [14]. In our case, radical

cystectomy was decided following the tissue diagnosis. No additional steps for treatment could be taken after radical cystectomy since the patient died before being discharged.

In conclusion, we think that clinicians should consider rare biphasic tumors such as sarcomatoid carcinomas of the bladder in patients presenting with hematuria in the differential diagnosis and a multidisciplinary treatment approach should be adopted.

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